Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/jcot

Case Report

Neurilemmoma of median nerve

Rammohan R M.B.B.S., M.S. (Ortho)^{a,*},
 Prince Gupta M.B.B.S., M.S. (Ortho)^a, Lalit Maini M.B.B.S., M.S. (Ortho)^b,
 Virender K. Gautam M.B.B.S., M.S. (Ortho), D.N.B., MNAMS^c

^a Senior Resident, Maulana Azad Medical College and Associated Lok Nayak Hospital, New Delhi, India

^b Professor, Maulana Azad Medical College and Associated Lok Nayak Hospital, New Delhi, India

^c Director Professor, Maulana Azad Medical College and Associated Lok Nayak Hospital, New Delhi, India

ARTICLE INFO

Article history:

Received 7 January 2014

Accepted 10 February 2014

Available online 30 March 2014

Keywords:

Schwannoma

Neurofibroma

Median nerve

Neurilemmoma

Nerve sheath tumors

ABSTRACT

Neurilemmomas constitute one of the most common tumors of peripheral nerves. Rarer amongst them is their occurrence in median nerve in the region of arm. A sixteen-year-old female presented with painless mass in right arm which was non-tender on palpation with positive Tinel's sign and no motor or sensory deficit in the affected arm. Ultrasound examination revealed an eccentrically arising mass of size 19 × 11 mm along median nerve. MRI study revealed a mass homogeneously isointense on T1 weighted images and hyperintense in T2 weighted images placed eccentrically in relation to median nerve in arm. Excisional biopsy under loupe magnification was carried out which revealed the mass to be neurilemmoma. This slowly growing benign tumor of peripheral nerves with an incidence of 5% with 14% involvement of Median nerve can be enucleated from the nerve with little or no damage. In spite of advanced imaging studies the mass cannot be differentiated preoperatively from another peripheral nerve sheath tumor neurofibroma. Both these tumors although bearing some clinical and imaging resemblance carry different intra-operative findings, histopathological features and post-operative results.

Copyright © 2014, Delhi Orthopaedic Association. All rights reserved.

1. Introduction

Neurilemmoma is a benign peripheral nerve tumor presenting as a painless mass in the extremity causing no or very little discomfort for the patient. Though rare, benign neurilemmomas are the most common tumors of the peripheral nerves.^{1,2} Differentiating from the neurofibroma is difficult preoperatively. Ultrasound and MRI if not diagnostic is helpful and suggestive in most of the cases. Treatment entails simple

excision which involves meticulous dissection from the fascicles of nerve bundle using magnification. As in most of the reported cases post-operative recovery is uneventful.

2. Case report

A sixteen-year-old right hand-dominant female presented with complaint of painless mass in right arm which slowly increased in size over a one year period with tingling

* Corresponding author. No A 43, Ground floor, Firozshah Kotla, Vikram Nagar, New Delhi 110002, India. Tel.: +91 7838071012.

E-mail addresses: rammohanbangalore@gmail.com, dr_ammohan@outlook.com (Rammohan R).
<http://dx.doi.org/10.1016/j.jcot.2014.02.002>

0976-5662/Copyright © 2014, Delhi Orthopaedic Association. All rights reserved.

Fig. 1 – Ultrasound photograph of the tumor: hypoechoic encapsulated lesion along nerve.

sensation radiating to forearm and hand on application of pressure over the mass. On Physical examination, a firm non-tender mass was palpable in the distal one third of right arm over anteromedial aspect. Tinel's sign along the distribution of median nerve was positive on gentle tapping over the mass. Neurological examination revealed no motor or sensory deficits in median, ulnar and radial nerve distributions. Plain radiograph of right arm did not reveal any osseous or soft tissue lesion. Ultrasound examination with Multi frequency Linear probe revealed a fusiform hypoechoic well encapsulated soft tissue mass measuring 19 mm × 11 mm in continuity of median nerve along anteromedial surface of mid third arm (Fig. 1). The mass was found to be expanding the nerve in

bilenticular shape, but arising eccentrically. The acoustic enhancement was noted deep to the mass. Power Doppler study was negative for intranodal or peri-nodal vascularity. MRI study revealed an eccentrically located mass anterolaterally in relation to median nerve with clearly identifiable nerve proximal and distal to the mass homogeneously isointense on T1 weighted images and hyperintense in T2 weighted images (Figs. 2 and 3). A provisional diagnosis of neurilemmoma was made and written informed consent was obtained from the patient for the procedure of excision. The surgery was performed under loupe magnification. Intra-operatively the mass was present eccentrically within the epineurium of the median nerve. The epineurium was longitudinally incised and carefully the surrounding splayed fascicles were dissected free from the tumor. The tumor was enucleated free from the nerve and this was possible without dividing any fascicles (Figs. 4 and 5). A tumor of size 2.2 cm × 1.7 cm × 1.5 cm was resected. Histopathological examination confirmed the diagnosis as neurilemmoma. Post-operatively patient remained neurologically intact and made uneventful recovery.

3. Discussion

Neurilemmomas are benign nerve tumors that originate from the cells of the Schwann sheath and, thus, are also called schwannomas. It can arise from any nerve covered with a Schwann cell sheath, which include the cranial nerves (except for the optic and olfactory), the spinal nerves and the autonomic nervous system.³ They usually occur in patients aged

Fig. 2 – MR imaging of the nerve sheath tumor (sagittal and coronal section). a. T1 weighted image sagittal section: Isointense mass along median nerve b. T2 weighted image sagittal section: hyperintense mass along median nerve c. T1 weighted image coronal section: isointense mass along median nerve.

Fig. 3 – MR imaging of the tumor (axial sections) a. T2 weighted axial section: hyperintense mass anterolateral to nerve b. T1 weighted axial section: isointense mass anterolateral to nerve.

Fig. 4 – Intraoperative photographs. Eccentrically placed tumor along the course of the nerve.

Fig. 5 – Intraoperative photographs. Tumor dissected free from tumor without dividing any fascicles.

30–60 years and have no race or sex predilection.^{4–6} Neurilemmomas most commonly occur in the head and neck involving the brachial plexus and spinal nerves with the upper and lower limbs being affected less often.⁷ They are mostly located on the flexor surfaces of the extremities, and the upper extremities are twice as likely to be involved as the lower limbs.^{5,8} Neurilemmomas constitute 5% of all benign soft tissue tumors,⁹ and are the most common benign nerve tumor occurring in the upper extremity.¹⁰ Rockwell et al reported 14% incidence of median nerve neurilemmoma.¹¹ Neurilemmomas usually grow slowly and appear as painless swelling for several years before being diagnosed.^{8,12,13} The onset of symptoms is usually associated with location rather than size of the tumor as symptoms of neural compression arise with growth of the mass.¹⁴ Spectrum of clinical presentation, in decreasing order, include palpable mass, Tinel's sign, paraesthesia, local tenderness, spontaneous pain, hyphoesthesia, rapid increase in size, incidentally detected mass and motor weakness.¹⁵ These present as swellings which are only painful to pressure and are mobile from side to side, but not in the vertical axis of the limb. Percussion induces painful paresthesiae in the area of the nerve of origin similar to Tinel's sign. It is suggested that this finding is the single most useful sign in the diagnosis of a neurilemmoma.¹⁶ The infrequency of neurilemmomas in general clinical practice causes difficulties in diagnosis and delay before recognition.¹⁷ Because these tumors are slow growing, soft in consistency, mobile in nature, and sometimes painless, they are sometimes misdiagnosed as lipoma, fibroma, ganglion or xanthoma.^{4,12} MRI is the investigation of choice to confirm the presence of a neurilemmoma.⁷ Typical features of a schwannoma on MR imaging are slightly hyperintense or isointense signal relative to skeletal muscle on T1 weighted images and increased, slightly heterogenous signal intensity on T2 weighted images.¹⁸ On fluid sensitive sequences the lesions may exhibit high signal intensity in the periphery and low to intermediate signal intensity centrally. This appearance has been termed the 'target sign' and is caused by the zonal architecture of the lesion with more myxoid material peripherally and more fibrous tissue centrally.¹⁹ Although ultrasonography is not 100% accurate in differentiating neural tumors from other tumors, or differentiating different kinds of neural tumors from each other, it is cheap and quick technique that can be used as an initial imaging modality.²⁰ Neurilemmomas appear as sharply delineated hypoechoic and homogenous masses with posterior acoustic enhancement sonographically.²¹ Because the recurrence and malignant transformation rates of neurilemmoma are low, simple removal of the tumor after careful dissection is recommended.^{4,22,23} With careful surgical technique and magnification, a neurilemmoma can be removed from the nerve sheath proper without creating any neurological deficit.²⁴ It has been found that the risk of developing neurological deficits were more likely to be high in patients with larger tumors and tumors associated with longer history.^{25,26} It was observed that larger tumors tended to have more fascicles entering the tumor substance and were at greater risk of major neurological deficits after surgery.²⁷ Schwannomas characteristically form an eccentric oval swelling, less than 3 cm in diameter, with the fascicles of the parent nerve

stretched and displaced over the dome of the mass.¹⁵ A plane is developed between the nerve bundles and the capsule of the tumor which is then removed whole.⁴ The neurilemmoma is enveloped by a true capsule which consists of the perineurium of the nerve bundle of origin, surrounded by a condensation of the deepest layers of the epineurium. On histopathological examination, the tumor contains varying proportions of two distinctive tissues, Antoni A and Antoni B. Antoni A are cellular areas with compactly arranged spindle cells frequently arranged in interlacing fascicles, palisades, with two compact parallel rows of well-aligned nuclei forming eosinophilic structures known also as Verocay bodies. Antoni B on the other hand, exhibits hypocellular areas consisting of a few tumor cells in loose myxomatous matrix lacking any distinctive architectural features.²⁸ A foci of inflammatory cells including histiocytes may also be present. Immunostaining for S100 protein is nearly always positive in the spindle cells of neurilemmomas, but negative or only weakly positive in most neurofibromas.²⁹ The neoplastic cells also express vimentin and myelin basic protein. Recurrence of schwannoma after surgical excision is rare.^{5,22,30,31} Recurrence has been seen in different areas of the nerves of the same extremity, but not in operated sites.⁶

4. Conclusion

Neurilemmomas, commonest amongst the peripheral nerve tumors, are amenable to enucleation without damaging the fascicles of the involved nerve. Advanced imaging studies cannot rule out the possibility of Neurofibroma and prudence dictates that a differential diagnosis of both these are kept prior to surgery.

Conflicts of interest

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

REFERENCES

1. Weiss SW, Goldblum JR, Enzinger FM. Benign tumours of the peripheral nerves. In: Weiss S, Goldblum JR, eds. *Enzinger and Weiss's Soft Tissue Tumours*. 4th ed. St. Louis: Mosby; 2001:1111–1208.
2. Giannini C. Tumors and tumor-like conditions of peripheral nerves. In: Dyck PJ, Thomas PK, eds. *Peripheral Neuropathy*. 4th ed. Philadelphia: Elsevier Saunders; 2005:2585–2606.
3. Zachariades N, Mezitis M, Vairaktaris E, et al. Benign neurogenic tumors of the oral cavity. *Int J Oral Maxillofac Surg*. 1987 Feb;16(1):70–76.
4. Kang HJ, Shin SJ, Kang ES. Schwannomas of the upper extremity. *J Hand Surg Br*. 2000;25:604–607.
5. White NB. Neurilemmomas of the extremities. *J Bone Joint Surg Am*. 1967;49:1605–1610.
6. Artico M, Cervoni L, Wierzbicki V, D'Andrea V, Nucci F. Benign neural sheath tumours of major nerves: characteristics in 119 surgical cases. *Acta Neurochir (Wien)*. 1997;139:1108–1116.

7. Nawabi DH, Sinisi M. Schwannoma of the posterior tibial nerve: the problem of delay in diagnosis. *J Bone Joint Surg Br.* 2007 Jun;89(6):814–816.
8. Holdsworth BJ. Nerve tumours in the upper limb. A clinical review. *J Hand Surg Br.* 1985;10:236–238.
9. Beaman FD, Kransdorf MJ, Menke DM. Schwannoma: radiologic-pathologic correlation. *Radiographics.* 2004 Sep–Oct;24(5):1477–1481.
10. Athanasian EA. Bone and soft tissue tumors. In: Wolfe SW, Hotchkiss RN, Pederson WC, Kozin SH, eds. *Green's Operative Hand Surgery*. 6th ed. Philadelphia: Elsevier Churchill Livingstone; 2011.
11. Rockwell GM, Thoma A, Salama S. Schwannoma of the hand and wrist. *Plast Reconstr Surg.* 2003;111:1227–1232.
12. Hecht OA, Hass A. Regional multiplicity of a neurilemmoma. *Hand.* 1982;14:97–99.
13. Kececi Y, Gurler T, Gundogan H, Bilkay U, Cagdas A. Benign giant schwannoma located in the upper arm. *Ann Plast Surg.* 1997;39:100–102.
14. Ozdemir O, Kurt C, Coskunol E, Calli I. Schwannomas of the hand and wrist: long-term results and review of the literature. *J Orthop Surg.* 2005;13(3):267–272.
15. Kim Sang-Min, Seo Sung-Wook, Lee Jun-Young, Sung Ki-Sun. Surgical outcome of schwannomas arising from major peripheral nerves in the lower limb. *Int Orthop (SICOT).* 2012;36:1721–1725.
16. Knight DM, Birch R, Pringle J. Benign solitary schwannomas: a review of 234 cases. *J Bone Joint Surg Br.* 2007 Mar;89(3):382–387.
17. Kehoe NJ, Reid RP, Semple JC. Solitary benign peripheral-nerve tumours: review of 32 years' experience. *J Bone Joint Surg Br.* 1995;77-B:497–500.
18. Hems TE, Burge PD, Wilson DJ. The role of magnetic resonance imaging in the management of peripheral nerve tumours. *J Hand Surg Br.* 1997;22:57–60.
19. Banks KP. Signs in imaging. The target sign: extremity. *Radiology.* 2005;234(3):899–900.
20. Ozdemir O, Ozsoy MH, Kurt C, Coskunol E, Calli I. Schwannomas of the hand and wrist: long-term results and review of the literature. *J Orthop Surg (Hong Kong).* 2005 Dec;13(3):267–272.
21. Simonovsky V. Peripheral nerve schwannoma preoperatively diagnosed by sonography: report of three cases and discussion. *Eur J Radiol.* 1997;25:47–51.
22. Phalen GS. Neurilemmomas of the forearm and hand. *Clin Orthop Relat Res.* 1976;114:219–222.
23. Whitaker WG, Droulias C. Benign encapsulated neurilemmoma: a report of 76 cases. *Am Surg.* 1976;42:675–678.
24. Louis DS, Hankin FM. Benign nerve tumors of the upper extremity. *Bull N Y Acad Med.* 1985 Sep;61(7):611–620.
25. Oberle J, Kahamba J, Richter HP. Peripheral nerve schwannomas – an analysis of 16 patients. *Acta Neurochir (Wien).* 1997;139:949–953.
26. Ogose A, Hotta T, Morita T, Otsuka H, Hirata Y. Multiple schwannomas in the peripheral nerves. *J Bone Joint Surg Br.* 1998;80:657–661.
27. Park MJ, Seo KN, Kang HJ. Neurological deficit after surgical enucleation of schwannomas of the upper limb. *J Bone Joint Surg Br.* 2009;91:1482–1486.
28. Arcot R, Ramakrishnan K, Rao S. Peripheral and cranial nerve sheath tumors-a clinical spectrum. *Indian J Surg.* 2012 Oct;74(5):371–375. <http://dx.doi.org/10.1007/s12262-011-0392-5>. Epub 2012 Jan 10.
29. Weiss SW, Langloss JM, Enzinger FM. Value of S-100 protein in the diagnosis of soft tissue tumors with particular reference to benign and malignant Schwann cell tumors. *Lab Invest.* 1983;49:299–308.
30. Donner TR, Voorhies RM, Kline DG. Neural sheath tumors of major nerves. *J Neurosurg.* 1994;81:362–373.
31. Rosberg HE, Ekerot L. Multifocal neurilemmomas in the same upper extremity. Case report. *Scand J Plast Reconstr Surg Hand Surg.* 1996;30:153–156.